

Meningioma - A Borderline Diagnosis

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Abstract

Sphenoidal wing meningioma is a slow-growing tumor originating in arachnoid epithelial cells. Solving such cases requires close cooperation between the neurosurgeon and the neuro-ophthalmologist. Material and method: We present the case of a 62-year-old patient who came to the ophthalmology clinic accusing the decrease of painless visual acuity in the right eye of about 4 months, with progressive evolution associated with headache, narrowing of the visual field in the temporal half, balance disorders, weight loss. The patient states that he lost his visual acuity in his left eye a year ago after repeatedly accusing the decrease in visual acuity in his left eye and for which he was recommended periodic eye evaluation for cataract surgery (at 6 months from the first visit the patient loses light perception in the left eye). Clinical examination establishes the diagnosis of LE Optic nerve atrophy and raises suspicion of RE Compressive optic neuropathy. Following paraclinical investigations, the diagnosis of bulky left sphenoid wing meningioma with frontal and temporal cerebral edema is established. Treatment with acetazolamide and mannitol is administered then the patient is directed to the Neurosurgery clinic where the tumor is excised by extended paramedian frontal pteral approach with cranialization of the frontal sinus. Preoperatively, the patient's informed consent is obtained – visual acuity in both eyes, without light perception. The anatomo-pathological examination diagnoses grade III anaplastic meningioma. Postoperatively, the patient performed MRI control and oncological treatment (radiation by intensity-modulated radiotherapy technique). Results: Immediately postoperatively the patient loses visual acuity in the right eye (without light perception), being warned by the surgeon in this regard, but the general condition is good. Conclusions: Early diagnosis would have been associated with a favorable visual and vital prognosis on both short and medium term. The management is multidisciplinary (close cooperation between neuro-ophthalmology and neurosurgery) and aims to optimize the patient's quality of life.

Keywords: meningioma, compressive optic neuropathy, optic atrophy

Introduction.

Sphenoidal wing meningioma is a slow-growing tumor originating in arachnoid epithelial cells. (1) It represents about 11-20% of all meningiomas. It affects women 3-6 times more than men, aged 40-60 years. The average age of onset is 63 years. (2) The incidence of meningiomas increases steadily after this age. The WHO (World Health Organization Classification of Tumors of The Central Nervous System) of brain tumors divides meningiomas clinically and anatomically-pathologically (3).

Grade I- secretory, microcystic with clear lymphoplasmocytic cells meningiomas. Despite the invasion of bone structures, grade I meningioma does not invade the brain parenchyma.

Grade II – atypical meningiomas (frequent mitosis and increased nucleus / cytoplasm ratio).

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Grade III – malignant, papillary and anaplastic meningiomas (intense mitosis, necrosis, invasion of the cerebral parenchyma).

The major risk factors for a more advanced WHO class are: age over 65 years and meningiomas that do not involve the cranial floor. Males were associated with twice the risk of advanced forms according to the WHO. (4)

Case report.

We present the case of a 62-year-old male patient who complains of vision issues in the right eye (RE) with 4 months onset before the presentation, with progressive evolution, painless, positive scotoma, headache, balance disorders, weight loss. The patient states that he has lost visual acuity in his left eye (AVOS) for about 1 year. In 2018, the patient performs various ophthalmological examinations to investigate the decrease in AVOS and periodic reassessment is recommended for the surgical

treatment of cataracts in the left eye (LE). 6 months after the first AVOS presentation = no light perception (FPL). AVOD 1 fc, AVOS FPL. Intraocular pressure AO 15 mmHg. The examination of the anterior segment highlights a round, central, reflex pupil RE, LE pupil in areflex semimidriasis, fine AO cortical crystalline opacities, otherwise normal appearance. The evaluation of the chromatic sense reveals RE unsystematized dyschromatopsia. The examination of the fundus reveals RE of the optic disc with clear contour, discreetly pale in the temporal sector, pallor exceeds the excavation, slightly dilated veins, narrowed arteries, microaneurysms and hemorrhages in the macular area, LE optic disc with clear contour, alpha and beta peripapillary atrophy, pale diffuse, more highlighted in the temporal sector, dilated veins, narrowed arteries, no lesions of the macula. (Fig. 1)



Fig. 1. The examination of the fundus

The perimeter of Humphrey 30-2 RE highlights the shrinking of the visual field in the temporal sector as right hemianopsia. (Fig. 2)

Optical nerve tomography results in a slight RE thin of the nerve fiber layer in the temporal sector, and at the LE level the nerve fiber layer (RNFL) have a moderate thinness in the lower sector and a significant thinning in the upper and temporal sectors (Fig. 3.)

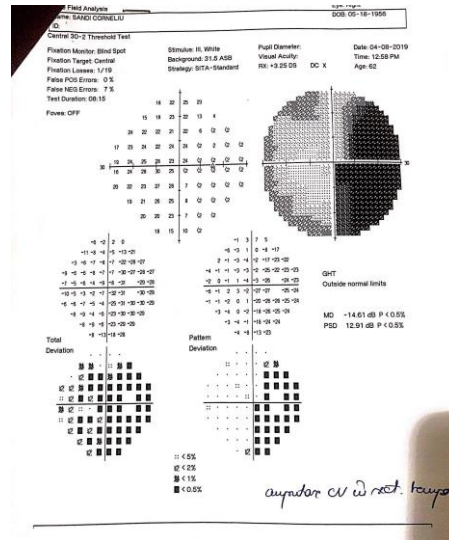


Fig. 2. OD CV 30-2 – visual field shrinking in the temporal sector of the right hemianopsia type

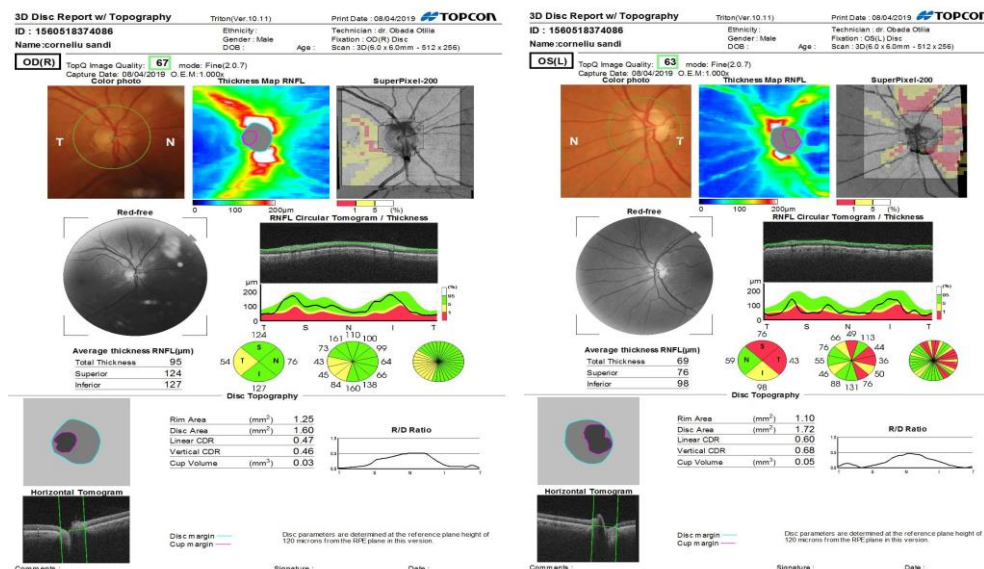


Fig. 3. OCT of optic nerve. Moderately thin RNFL RE in the temporal sector, moderately thin RNFL LE in the lower sector and important in the upper and temporal sectors

The biological balance shows hyperglycemia and hypercholesterolemia.

The emergency imaging examination (craniocerebral and orbit CT with contrast substance)(under the close monitoring of the collaborating anesthetist, considering the influenced condition of the patient) (Fig. 4) shows a solid expansive formation with intrinsic millimeter calcifications, with significant contrast, located left fronto-temporal extranevax, plated with sphenoid bone with overall dimensions of 66/70/30 mm, showing the following ratios:

- it circumscribes the left cavernous sinus and extends to the pituitary gland,
- it extends posteriorly from the clivus to a length of ~ 20mm,
- has a mass effect on the midbrain,
- has a close relationship with the basilar trunk, without changes in caliber.
- the supracondylar segment of the left internal carotid artery is involved

- laterally it extends along the meninges at the level of the middle fossa, it includes the sphenoparietal sinus.
- on the upper side it has a mass effect on the lower face of the left frontal lobe.
- The optical chiasm is not identified
- Diffuse edema of the white matter at the left frontal and temporal level that causes subfalciform hernia with displacement to the right ~ 7mm.

Being in front of a neurosurgical emergency, it is decided to administer the emergency treatment (Manitol, Acetazolamide) and to direct the patient to the neurosurgery service.

Within the neurosurgery service, craniocerebral MRI is performed (Fig.5): extraaxial formation plated on the left sphenoidal wing, extending to the suprasellar, prepontine cisterns and in the anterior floor of the skull base; of approximately 70/80/50 mm (AP / TT / CC). At the intraslesional level it contains small calcifications and several vessels, some with rapid flow. It protrudes

posteriorly in the suprasellar cistern, in the Turkish saddle, compresses the pituitary gland, invades both cavernous sinuses, predominantly on the left one; gets in contact with the orbital apexes, the chiasm is also embedded in the nerves III, IV, V; compresses the left olfactory bulb, protrudes into the left upper ethmoid cells. It exerts a mass effect on the proximal vascular structures in the vicinity, without changing their permeability.

Following the performed investigations, the diagnosis is of LE Compressive optic neuropathy, Voluminous meningioma of the left sphenoidal wing.

Under the patient's preoperative informed consent (AVAO FPL), surgery is performed and the macroscopic, subtotal resection with decompression of both optic nerves is performed. AVAO FPL postoperative. The anatomopathological examination revealed grade III anaplastic meningioma.

Postoperatively, the patient is re-evaluated and directed to the oncology service for brain irradiation by the IMRT technique DT = 51Gy/30fr/PTV and the treatment of related diseases.

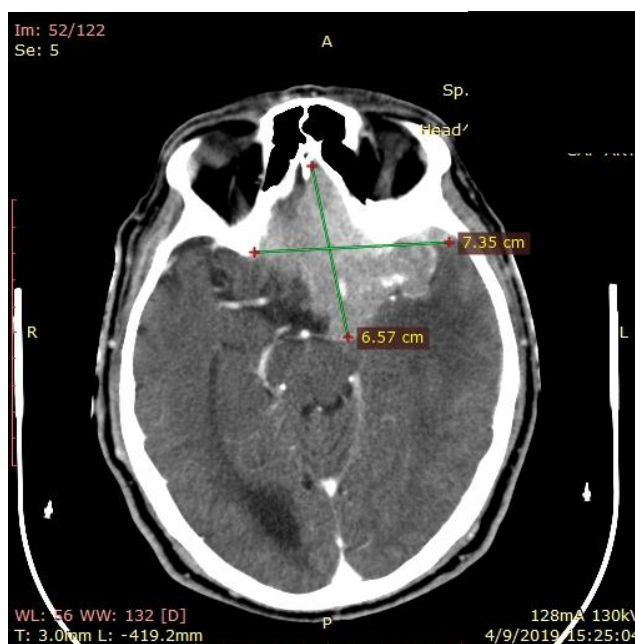


Fig. 4. Craniocerebral CT with contrast substance examination



Fig. 5. Craniocerebral MRI examination

At 4 months postoperatively the patient is re-evaluated ophthalmologically. AVAO FPL. The examination of the anterior pole: AO pupils in mydriasis, areflex. The

examination of the fundus reveals a completely pale AO optical disc, more stressed in the temporal sector (Fig. 6). OCT shows the emphasis of the nerve fiber layer thinning (Fig. 7).



Fig. 6. Fundus examination at 4 months postoperatively: AO completely pale optical disc, more stressed in the temporal sector

The peculiarity of the case consists in the late diagnosis of the primary cause that determined the decrease of visual acuity, when it jeopardized the patient's life, AO FPL postoperative visual acuity, subtotal resection and the need for postoperative radiotherapy. The interdisciplinary cooperation is also crucial, in this case with specialized

doctors in neurosurgery. The medium and long term visual prognosis is reserved.

The medium-term vital prognosis is favorable but the long-term one is reserved.

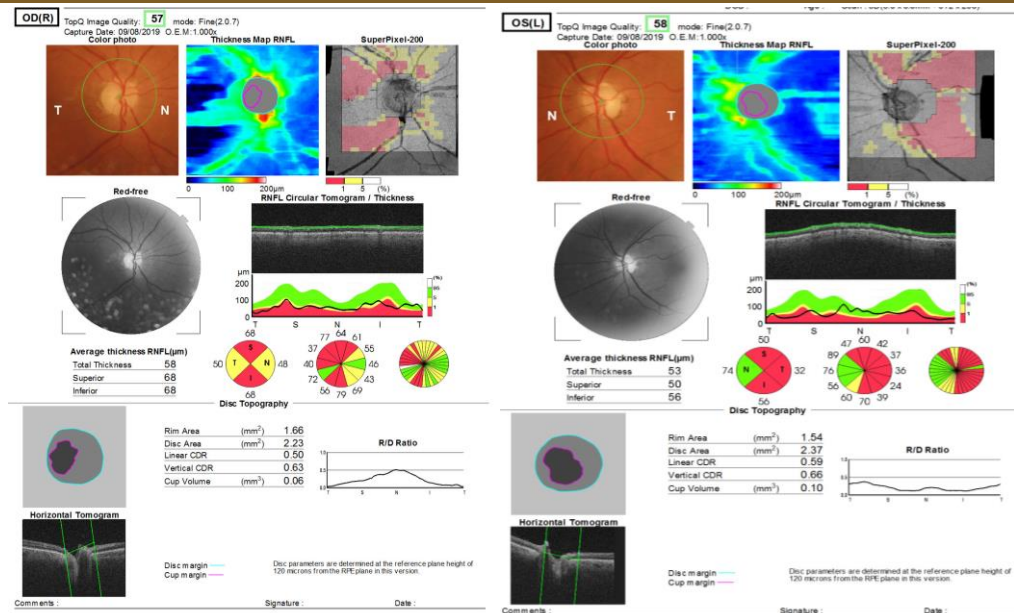


Fig. 7 OCT optic nerve examination at 4 months postoperatively

Discussions.

Sphenoidal wing meningioma is a slow-growing tumor originating in the arachnoid epithelial cells. (1)

The anterior skull base meningioma is 40% of all intracranial meningiomas. (5) Sphenoidal wing meningioma represents approximately 11-20% of all meningiomas. It affects women 3-6 times more than men, aged 40-60 years. The average age of onset is 63 years. (2) The incidence of meningiomas increases steadily after this age. Based on clinical observations and anatomical features, sphenoid meningiomas are divided into: internal third tumors of the sphenoid wing, mid-ridge tumors, "en plaque" pterial tumors and global pterial tumors. Medial localized meningiomas are accompanied by morbidity, mortality, and a higher recurrence rate compared to other forms of meningioma because they involve the anterior localized intracranial arteries, the anterior segment of the visual pathway, and the cavernous sinus. (6) The WHO (World Health Organization

Classification of Tumors of the Central Nervous System) brain tumors divide meningiomas clinically and pathologically into:

Grade I - secretory, microcystic with clear lymphoplasmocytic cells meningiomas. Despite the invasion of bone structures, grade I meningioma does not invade the brain parenchyma.

Grade II - atypical meningiomas (frequent mitosis and increased nucleus / cytoplasm ratio).

Grade III - malignant, papillary and anaplastic meningiomas (intense mitosis, necrosis, invasion of the brain parenchyma).

The growth rate of meningiomas is on average 1-3 mm per year. (7) Malignant meningiomas (WHO Grade III) are characterized by an increased number of intracellular abnormalities but also a higher growth rate than benign or atypical meningiomas. Also, both the invasion and recurrence rates of the brain are higher compared to the other two subtypes. (8)

A retrospective study that included 1663 patients subjected to surgery for meningioma revealed that 90% of tumors were benign (WHO grade I), while only 10% were malignant (WHO grade II-III). The major risk factors for a more advanced WHO class are: age over 65 years and meningiomas that do not involve the skull base. Males were associated with twice the risk of advanced types according to the WHO. The secondary morbidity of the tumor or of the therapeutic interventions depends on its location but also on its proximity to certain ocular structures or vital neurological areas. (4) No risk factors were identified. Genetically, it is associated with the absence of the NF2 gene on the long arm of chromosome 22 (22q), which encodes merlin-a tumor suppressor. 60% of meningiomas are associated with this mutation. They may also be associated with Gorlin or Rubinstein-Taybi syndromes, but the association is not as close as that of the NF2 gene mutation. Meningiomas can be multiple, particularly when associated with type 2 neurofibromatosis. (9)

The diagnosis will take into account a number of important aspects: impairment of vision is correlated with the location of the tumor with conal or extraconal touch, this being associated with more severe impairment of visual acuity; optic nerve compression occurs late and is correlated with compression of the intraorbital side of the

optic nerve; visual field: temporal hemianopsia / central scotoma / peripheral narrowing / central superior scotoma. If the dura mater is also involved, the patient may complain of pain. (10) The literature describes cases in which the first manifestation of a sphenoidal wing meningioma was depression or behavioral disorders (11), as well as acute intracerebral haemorrhage (12) or hearing loss (13). Due to the rarity of this tumor, only a few studies have been published on the surgical treatment of sphenoid meningioma in the literature. Complete surgical resection is difficult due to the involvement of the orbital apex, the superior orbital fissure and the cavernous sinus, so it has higher recurrence rates. The treatment of meningioma depends on several factors such as: the size and location of the tumor, the type of meningioma and age. The surgery is difficult to perform due to the location and difficulty of the approach requiring close cooperation with the neurosurgeon, anesthetist and the ophthalmologist. Postoperative evaluation may reveal the degree of ocular damage following partial or total resection of the tumor, visual, functional and vital prognosis. (2,3). According to the WHO classification, the patient was diagnosed with grade III meningioma (anaplastic meningioma).

The slow, unilateral, painless decrease in the patient's visual acuity in the LE to AVOS FPL without other changes that would suggest an intraorbital injury or that could be explained by changes in the anterior pole, should lead us to think of an expansive intracranial formation that causes secondary compressive optic neuropathy. As mentioned, meningiomas are tumors with a slow growth of about 1-3 mm per year. (1, 7) However, in the present case we are talking about a malignant meningioma that is characterized by a faster growth rate compared to the other two subtypes. (8) If we take into account this, but also the fact that visual disturbances in the LE began long before the RE, we think that the visual and vital prognosis of the patient would have been perhaps better in terms of early diagnosis.

Conclusions.

Early diagnosis would have been associated with a favourable visual and vital prognosis on both short and medium term. The management is multidisciplinary (close cooperation between neuroophthalmos, neurosurgery, and anesthesia intensive care) and aims to optimize the quality of life of the patient.

Conflicts of interest

The authors do not declare any conflicts of interest.

Authors' contributions

The authors have read and approved the final version of this article.

References

1. Basic and Clinical Science Course. Orbit, Eyelids, and Lacrimal System. American Academy of Ophthalmology. 2019-2020
2. Mubeen B, Makhdoomi R, Nayil K, et al. Clinicopathological Characteristics of Meningiomas: Experience from a Tertiary Care Hospital in the Kashmir Valley. *Asian J Neurosurg*. 2019;14(1):41-46.
3. Louis, D.N., Perry, A., Reifenberger, G. et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol* 131, 803–820 (2016).
4. Buerki RA, Horbinski CM, Kruser T, Horowitz PM, James CD, Lukas RV. An overview of meningiomas. *Future Oncol*. 2018;14(21):2161-2177.
5. Güdük, M., Özdoğan, K. & Pamir, M. N. Sphenoid Wing Meningiomas: Surgical Outcomes in a Series of 141 Cases and Proposal of a Scoring System Predicting Extent of Resection. *World Neurosurgery* 125, e48–e59 (2019).
6. Nakamura, M., Roser, F., Jacobs, C., Vorkapic, P. & Samii, M. Medial Sphenoid Wing Meningiomas: Clinical Outcome and Recurrence Rate. *Neurosurgery* 58, 626–639 (2006).
7. Sherman, et al. Chemotherapy: what is its role in meningioma? *Expert Rev. Neurother*. 12(10), 1189–1196 (2012)
8. Garzon-Muvdi, T., Yang, W., Lim, M. *et al*. Atypical and anaplastic meningioma: outcomes in a population based study. *J Neurooncol* 133, 321–330 (2017).
9. Lee YS, Lee YS. Molecular characteristics of meningiomas. *J Pathol Transl Med*. 2020;54(1):45-63.
10. Hussaini SM, Dziurzynski K, Fratzkin JD, Jordan JR, Hussain SA, Khan M. Intraosseous meningioma of the sphenoid bone. *Radiol Case Rep*. 2015;5(1):357.
11. Ciobanu AM, Lisievici M.Gh, Coman TC, Ciubotaru V.Gh, Drăghia A., Ciucu A.A. Giant wing sphenoid meningioma with principal manifestation depression, *Roumanian Journal Of Morphology and Embryology* 2009, 50(4): 713-717.
12. Frič R, Hald JK, Antal EA. Benign Sphenoid Wing Meningioma Presenting with an Acute Intracerebral Hemorrhage - A Case Report. *J Cent Nerv Syst Dis*. 2016;8:1-4.
13. Del Toro E, Risbud A, Khosravani N, Vengerovich G, Archilla A. Sphenoid Wing Meningioma Presenting as Sudden Sensorineural Hearing Loss: A Case Report and Literature Review. *Ear Nose Throat J*. Feb 2020.